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ORIGINAL ARTICLE

Medical Certificates of Cause of Death for people with intellectual disabilities: A systematic literature review

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The Learning Disabilities Mortality Review (LeDeR) Programme is commissioned by the Healthcare Quality Improvement Partnership (HQIP) on behalf of NHS England.

Background: Mortality studies can help reduce health inequalities by informing public policy through a better understanding of causes of death and comorbidities. Mortality studies often rely on Medical Certificates of Cause of Death (MCCD) for data.

Method: A systematic review was undertaken to identify the extent and nature of issues in recording causes of death for people with intellectual disability on MCCD.

Results: Fifteen of the 25 articles included in the literature review raised concerns about the accuracy of MCCD in identifying the cause(s) of death of people with intellectual disability. The most frequent issues were the under-reporting of intellectual disability on MCCD, and listing intellectual disability or an associated condition as an underlying cause of death.

Conclusions: Concerns about the accuracy and reliability of MCCD for people with intellectual disability raise questions about mortality data based on MCCD. Clear guidance is required from WHO for those completing MCCD for people with intellectual disability.

KEYWORDS

intellectual disabilities, Medical Certificate of Cause of Death, mortality

1 | INTRODUCTION

Mortality studies are an established resource for research into trends in the health of a population (WHO, 2014). They allow for comparisons between and within populations and are a method for analysing the success of health policies and interventions. As stated by Jha (2012), “counting the dead is one of the world’s best investments to reduce premature mortality worldwide as it is one of the most robust ways to measure accurately the effectiveness of investments aimed at reducing ... mortality” (p. 1). Accurate and consistent reporting of causes of death is critical for creating reliable data to be used by mortality studies. Highlighting the importance of mortality studies, Swain, Ward, and Hartlaub (2005) note,

“the primary tool for measuring mortality rates is the death certificate” (p. 652).

In 1927, the World Health Organization (WHO) recommended the Medical Certificate of Cause of Death (MCCD) to be the format to record the cause of a person’s death. The MCCD is divided into two sections, parts 1 and 2. Contained in Part 1 is the immediate cause of death, tracking the sequence of causes back to any underlying cause or causes. The WHO defines the underlying cause of death as “a) the disease or injury which initiated the train of morbid events leading directly to death, or b) the circumstances of the accident or violence which produced the fatal injury” (WHO, 2010, p. 31). Part 2 of the MCCD is used to list other significant conditions, diseases or injuries that contributed to the death, but were not part

of the direct sequence leading to death. The MCCD is completed by a medical practitioner or in some cases a coroner.¹ The information from the MCCD is then coded using the latest International Classification of Disease (ICD) codes, to form national statistics on the causes of death of a population.

The quality and reliability of MCCD for use in mortality studies is a source of scrutiny and debate: studies in Australia, Chile, Estonia, Sri Lanka, Greece, the United Kingdom and the United States have noted common problems as being ill-defined causes of death or modes of death, improper causal sequences and the mechanism rather than the cause of death being reported (Antini, Rajs, Munoz-Quezada, Mondaca, & Heiss, 2015; Bell, Gaitatzis, Johnson, & Sander, 2004; Bugeja, Clapperton, Killian, Stephan, & Ozanne-Smith, 2010; Cheng, Lu, & Kawachi, 2012; Katsakiori, Panagiotopoulou, Sakellaropoulos, Papazafropoulou, & Kardara, 2007; Rahu, Palo, & Rahu, 2011; Rampatige, Gamage, Peiris, & Lopez, 2013).

There are several factors that may influence the accuracy and effectiveness of MCCD in general. The most commonly reported factor is insufficient training or a lack of knowledge about completing the MCCD on the part of the certifier (Bell et al., 2004; Harriss et al., 2011; Mahdavi, Sedghi, Sadoghi, & Fard Azar, 2015; Rampatige et al., 2013). Other factors associated with the accuracy and effectiveness of MCCD are the profession of the certifier (Katsakiori et al., 2007; Mahdavi et al., 2015; McKenzie, Chen, & Walker, 2009), the number of causes of death reported (Antini et al., 2015), a lack of knowledge of the medical history of the deceased person (Katsakiori et al., 2007), the frequency of use of autopsy in a country (Ylijoki-Sorensen et al., 2014) and multiple languages used within a country (Haghighi, Dehghani, Teshizi, & Mahmoodi, 2014). In addition, the frequency of the use of autopsy has seen a major decline over past decades for people regardless of disability status (Ministry of Justice 2016; Shojania & Burton, 2008), and concerning rates of poor quality autopsies have been reported (Kuijpers et al., 2014; NCEPOD 2006).

The accuracy of the MCCD in relation to people with intellectual disability is of key importance in understanding the excess mortality of this population (Florio & Trollor, 2015), offering guidance for planning policies and practices to reduce premature mortality (Heslop, Lauer, & Hoghton, 2015), and monitoring the effectiveness of such policies and practices (Lauer & McCallion, 2015). The reliability of MCCD in relation to people with intellectual disability has been called into question largely through individual-level mortality reviews which scrutinize the sequence of events leading to the death of an individual; these have noted discrepancies between narratives of how and why people have died, and their causes of deaths recorded on the MCCD (Heslop et al., 2014; Hollins, Attard, von Fraunhofer, McGuigan, & Sedgwick, 1998).

The objective of this study is to present the findings of a systematic review of research pertaining to the accuracy of MCCD for identifying causes of death of people with intellectual disability. The review summarizes research that identifies potential difficulties in relying on

MCCD to help understand the causes of death of people with intellectual disability, why these difficulties may occur and the impact they have. The study concludes with suggestions for strengthening reporting the causes of death of people with intellectual disability on MCCD.

2 | METHOD

Electronic literature database searches were conducted in PubMed, ProQuest, CINAHL (Cumulative Index for Nursing and Allied Health Literature) and Web of Science in July 2017. Depending on the functionality of the database, searches combined terms for mortality, intellectual disability and death certification. Hand searching (from reference lists of studies meeting the inclusion criteria) was carried out to retrieve additional sources. Articles included research, reviews or investigations into mortality of people with intellectual disability, with reference to MCCD. Specific inclusion and exclusion criteria were applied as follows:

Inclusion criteria

- Article in the English language
- Published during the past 20 years (1997–2017)
- Reporting quantitative, qualitative or mixed methods research, audit or evaluation
- Peer-reviewed articles or policy guidance documents
- Studies (including those that review the accuracy of MCCD) where results are disaggregated for people with intellectual disability
- Studies reporting trends in cause of death, age at death, avoidable death or comorbidities at death of people with intellectual disability

Exclusion criteria

- Reviews of studies, commentaries, editorials or abstracts from meetings or conferences
- Studies about the general population, or about people with specific syndromes where intellectual disability cannot be assumed, that do not disaggregate data for people with intellectual disability
- Studies about specific syndromes associated with intellectual disability with the exception of Down's syndrome (the most common genetic cause of intellectual disability)

The first author checked the titles and abstracts from the initial search to exclude studies that were obviously not in scope. Articles that were retained for a review of the full text were those that were potentially in scope or those about which a decision could not be made solely on the basis of the title and abstract. Following a review of the full text, and any additional hand searching of references or citations, articles meeting the inclusion criteria were summarized in tabular form recording the author(s) and year, country, study design, aim of the study, sample size and age range (see Tables 1 and 2). The articles are split into two tables on the basis of whether they identify issues relating to MCCD.

¹Coroners investigate deaths where the cause is unknown, where there is reason to think the death may not be due to natural causes, or which need an inquiry for some other reason.

3 | RESULTS

The summary of the process followed for identifying articles for inclusion is displayed in Figure 1.

Searches identified 133 articles. The titles and abstracts of the publications were read, and after eligibility criteria were applied and duplicates deleted 22 articles were retained. Hand searching reference lists and citations added six articles. Three articles were excluded after reading the full texts. A total of 25 articles form the basis for the findings of this literature review.

3.1 | Concerns about the accuracy of MCCD in identifying the cause of death of people with intellectual disability

The 25 articles identified in the review mostly relate to the United States and the UK; however, articles from Australia, Canada, Finland, Germany, India and Ireland are included. The 25 articles pertain to 24 studies. All are primary studies, apart from Heslop and Glover (2015), Heslop et al. (2014), McCarthy and O'Hara (2011), Ouellette-Kuntz (2005) and Ouellette-Kuntz, Shooshtari, Balogh, and Martens (2015) which summarize or synthesize the findings of other studies or of national data. No articles that reviewed the accuracy of MCCD and disaggregated data for people with intellectual disability were found. Of the 25 articles relating to mortality of people with intellectual disability, 15 raised concerns about the accuracy of MCCD in identifying the cause of death of people with intellectual disability. Within these studies, two key issues were identified as follows: first, the under-reporting of intellectual disability on the MCCD, and secondly, inappropriately listing intellectual disability or associated conditions as the underlying cause of death.

3.2 | Under-reporting of intellectual disability on the MCCD

The joint most consistent concern, reported in eight of the articles, was the under-reporting of intellectual disability or associated conditions on the MCCD. Failing to include on the MCCD that a person had intellectual disability where this may have been a contributory cause of death diminishes the effectiveness of using MCCD data for analysing mortality trends and patterns in people with intellectual disability. This is not a newly identified problem. In 1998, Hollins et al. found only 40% of MCCD of people with intellectual disability in a London borough appropriately referenced that the person had intellectual disability or an associated condition. More recent data suggest that the MCCD appropriately recorded that the person had intellectual disability in 41% of deaths of people with moderate or severe intellectual disability (Tyrer & McGrother, 2009); 58% of people with profound or multiple intellectual disability; and only 9% of people with mild intellectual disability (Heslop et al., 2014). Glover and

Ayub (2010) noted that some causes of intellectual disability, for example, Down's syndrome, were better reported than others on the MCCD, but conditions, such as Fragile X syndrome and autistic spectrum conditions, were particularly poorly recorded. Discussing the issue of under-reporting that a person had intellectual disability on the MCCD, Hosking et al. (2016) suggest that this "emphasizes the limitations of studies based on death certificates alone" (p. 1488).

3.3 | Listing an intellectual disability or an associated condition as an underlying cause of death

Identified in eight of the articles and the other joint most common concern was the recording of intellectual disability or an associated condition, as the underlying cause of death. Landes and Peek (2013) argue that although this was previously acceptable practice because intellectual disability "was historically considered a disease process" (p. 1183), it should now be considered incorrect because intellectual disability is more accurately described as a disability (p. 1184). This builds on the assertion of Tyrer and McGrother (2009) that intellectual disability should not be included in Part 1 of the MCCD as it predisposes the individual to a fatal condition and is not the cause of the fatal condition (p. 902). Thus, intellectual disability should more appropriately be recorded in Part 2 of the MCCD, not Part 1.

In two separate studies into mortality of children and young people with Down's syndrome in the United States, Down's syndrome was listed as the underlying cause of death in 21.6% (Goldman, Urbano, & Hodapp, 2011) and 21.2% (Miodrag et al. 2013) of deaths. Other more recent studies reporting intellectual disability or an associated condition listed as the underlying cause of death are Hosking et al. (2016) and Trollor, Srasuebku, Xu, and Howlett (2017). As noted by Trollor et al. (2017), this practice "obscures relevant and potentially avoidable causes of death for this population and should be formally revised" (p. 8).

3.4 | Explanations for the inaccuracies in recording the cause of death of people with intellectual disability on MCCD

Several articles propose explanations for the inaccuracies in recording the cause of death of people with intellectual disability on MCCD. Hollins et al. (1998) reported that the high level of coding error found in their study could be a result of the certifier being "on duty" at the time of the death but with little knowledge of the person's medical history, a proposal supported by Landes and Peek (2013). Landes and Peek (2013) explain the erroneous coding of "mental retardation" on 20% of 2,278 MCCD of people with intellectual disability in the United States as being due to "contextual factors" (p. 118) including: a lack of knowledge of the deceased person's medical history; the number of causes of death recorded on the MCCD; the place of death being an emergency room or other location where medical records may be difficult to access; and an accidental cause of death such as poisoning or injury.

TABLE 1 Articles that raised concerns about the accuracy of MCCD in identifying the cause of death of people with intellectual disability*

Authors and year	Country	Study aim	Study design	Sample size	Age range (years)
Dieckmann, Giovis, and Offergeld (2015)	Germany	Analyse age group-specific mortality rates and the average life expectancy of people with intellectual disabilities.	Age group-specific mortality rates were estimated by exponential regression analyses on samples from Westphalia-Lippe and Baden-Württemberg 2007–2009.	24,500	20–90
Florio and Trollor (2015)	Australia	Get age-adjusted death rates (ADRs), standardized mortality ratios (SMRs) and age-standardized death rates (ASDRs) for persons with an intellectual disability.	Longitudinal cohort study linking retrospective data June 2005 to 31 December 2011.	42,219	20–85
Glover and Ayub (2010)	England	Study the ages and causes of death for people with intellectual disabilities or conditions which can cause Intellectual disabilities, who died in England between 2004 and 2008.	Employing death certificates from the Office for National Statistics, they analysed the cause of death for conditions associated with intellectual disability and applied standardized mortality odds ratios (SMORs).	5,430	2–87
Goldman et al. (2011)	The United States	Examine the amount, timing and causes/correlates of infant mortality among newborns with Down's syndrome.	The Tennessee Department of Health Birth, Hospital Discharge and Death records, from 1990 to 2006, were used to compare three death groups and the survival group on correlates of mortality.	1,305	0–1
Heslop et al. (2014)	International	Review why an understanding of mortality data in relation to people with intellectual disabilities is an important area of concern.	Reviews existing consensus on trends and issues in mortality data among people with intellectual disabilities.	NA	NA
Heslop and Glover (2015)	England	Examine research into mortality in England to increase evidence base focusing on SMR and age at death.	Uses data from the Confidential Inquiry into premature deaths of people with learning disabilities, the 2013 Joint Health and Social Care Intellectual Disability Self-assessment Exercise, local registers of people with intellectual disability and MCCD.	NA	NA
Hollins et al. (1998)	England	Examine the risk of early death, causes of death and the quality of data recorded on death certificates.	Multivariate survival analysis (Cox regression) was carried out on data from Learning Disability Registers and death records in two London districts between 1982 and 1990.	2,000	5 to ≤100
Hosking et al. (2016)	England	Compare mortality among adults with intellectual disability in England with the general population.	A cohort study from 2009 to 2013 using death certificates linked with data from 343 general practices.	16,666	18–84
Kiani et al. (2014)	England	Study mortality from sudden unexpected death in epilepsy (SUDEP) in adults with intellectual disability.	Individuals with intellectual disability between 1993 and 2010 were identified using the Leicestershire Intellectual Disability Register database. Cases of probable and definite SUDEP in adults with intellectual disability were compared with the general population using standardized mortality ratios (SMRs).	898	20 to >50

(Continues)

TABLE 1 (Continued)

Authors and year	Country	Study aim	Study design	Sample size	Age range (years)
Landes and Peek (2013)	The United States	Explore the effects of diagnostic ambiguity on risk of "mental retardation" being coded as underlying cause of death on US death certificates from 2004.	Utilizing all US death certificates from 2004 that included a cause of death code for mental retardation, logistic regression analysis provided estimates of the likelihood of having mental retardation erroneously coded as the underlying cause of death.	2,278	21–103
Miodrag et al. (2013)	The United States	Examine mortality trends among children and young adults with Down syndrome in the United States.	Hospital discharge and death records from the state of Tennessee were linked to examine 2046 hospitalized individuals with Down syndrome.	2,046	1–29
Raitasuo et al. (1997)	Finland	Examine the causes of death of people with intellectual disabilities in a residential home, and we compared them to the causes of death of the general population.	The causes of death, contributing factors and associated diseases were collected from death certificates and patient documents. The causes of death were based on autopsy in 85% of the patients. The ratio of observed (O) and expected (E) cases was compared.	216	1–86
Trollor et al. (2017)	Australia	Investigate mortality and its causes in adults over the age of 20 years with intellectual disability.	Retrospective population-based standardized mortality of the intellectual disability and comparison cohorts 2005 to 2011 in New South Wales.	19,362	20+
Tyrer and McGrother (2009)	England	Explore cause-specific mortality in adults with intellectual disability compared with the general population.	Cause-specific standardized mortality ratios (SMRs) were calculated by age and sex for adults with moderate to profound intellectual disability living in the unitary authorities of Leicester, Leicestershire and Rutland, UK, between 1993 and 2006.	2,995	20–70+
Yang, Rasmussen, and Friedman (2002)	The United States	Study patterns of mortality and morbidity in people with Down's syndrome who died during a 15-year period. Assess changes in age at death by racial group, most frequent diseases associated with death and occurrence of major categories of malignant neoplasms.	Used data from US death certificates from 1983 to 1997 to calculate median age at death and standardized mortality odds ratios (SMORs) for common medical disorders in people with Down's syndrome.	17,897	1–70

*A source qualifies as considering Medical Certificates of Cause of Death (MCCD) as an issue if they specifically mention the use of MCCD as a limitation of their research; there is an impact on the research methods; for example, under-reporting of intellectual disability is found, or intellectual disability is listed as an underlying cause.

TABLE 2 Articles that did not raise concerns about the accuracy of Medical Certificates of Cause of Death in identifying the cause of death of people with intellectual disability

Authors and year	Country	Study aim	Study design	Sample size	Age range (years)
Arvio, Salokivi, Tiittinen, and Haataja (2016)	Finland	Calculate the standardized mortality ratios (SMR) for those with an intellectual disability.	KELA's statistical database (www.kela.fi/tilastot) used to find the number and age of individuals who received benefits relating to intellectual disability who died 1996–2011.	NA	NA
Heslop et al. (2014)	England	To provide evidence about contributory factors to avoidable and premature deaths in this population.	All known deaths of people with intellectual disability were reviewed in South West England between June 2010 and May 2012. A control group was used to compare mortality rates.	247	4–75
Hogg, Juhlberg, and Lambe (2007)	Scotland	Track the ongoing implementation of policies with respect to a cohort of people with profound intellectual and multiple disabilities, including mortality and its causes.	A 10-Year Longitudinal Study of People with Profound Intellectual and Multiple Disabilities using a questionnaire.	142	N/A
Lakhan and Kishore (2016)	India	Investigate the mortality rate in people with intellectual disability and its association with age in rural and urban settings in India.	Secondary data from a national survey upon which a t test was carried out.	70,302 households	N/A
Lauer and McCallion (2015)	The United States	Calculate average age at death and crude mortality rates for people with intellectual disabilities.	Data from both US state intellectual disabilities service system administrative data sets and state Medicaid claims were used to calculate SMRs.	13,809	N/A
McCarron, Carroll, Kelly, and McCallion (2015)	Ireland	Calculate for mortality rates in the general Irish population compared to those with intellectual disabilities 2003–2012.	A standardized mortality ratio (SMR) and an average age at death. Data were drawn from the 2012 National Intellectual Disability Database and the Census in Ireland.	31,943	0–80+
McCarthy and O'Hara (2011)	The United States	Review morbidities and life expectancy of people with intellectual disabilities.	Reviewed the evidence base published in the past 12–24 months prior.	N/A	N/A
Ouellette-Kuntz (2005)	Canada	Explore health of individuals with intellectual disabilities, including life expectancy and mortality.	A review paper combining the literature related to the concepts of inequity and inequality with the body of knowledge on health disparities faced by individuals with intellectual disabilities.	N/A	N/A
Ouellette-Kuntz et al. (2015)	Canada	Explore mortality among people with intellectual disabilities in Canada.	Hospital data for in-hospital mortality among adults with intellectual disabilities in Ontario were linked to a matched set in Manitoba.	N/A	N/A
Tyrer, Smith, and McGrother (2007)	England	Measure the extent of excess mortality in people with intellectual disability compared with the general population.	Indirectly standardized all-cause and disease mortality ratios (SMRs) and exact Poisson confidence intervals were calculated by age and sex for all adults, aged 20 years or over, with moderate to profound intellectual disability living in Leicestershire and Rutland, UK, between 1993 and 2005.	2,436	20–70+

"Diagnostic overshadowing" defined by Tyrer and McGrother (2009) as "erroneously attributing presenting health problems to intellectual disability rather than to the underlying condition" (p. 903) has been and persists as an issue in healthcare provision for people with intellectual disability (Disability Rights Commission 2006; Jopp & Keys, 2001; Landes & Peek, 2013; Reiss, Levitan, & Szyszko, 1982). Diagnostic overshadowing has also been identified as being a factor in the accuracy of MCCD, with Kiani et al. (2014) concluding that "people with ID continue to experience diagnostic overshadowing even after their death" (p. 517).

A further cause of problems identified in relation to MCCD of people with intellectual disability condition has its roots in the multiple possible ICD-10 codes for intellectual disability. In their study of causes of death of people with intellectual disability in England and Wales, Glover and Ayub (2010) report 48 ICD-10 codes for medical conditions commonly associated with intellectual disability and 76 ICD-10 codes for conditions less commonly associated with intellectual disability, leading to increased potential for coding error (Heslop et al., 2014).

A lack of training and knowledge on behalf of certifiers is a likely further cause of inaccuracies in recording the cause of death of people with intellectual disability on MCCD. While not a common

explanation identified in the review pertaining to people with intellectual disability, it is the most frequent reason given in relation to the accuracy of MCCD for the general population. Swain et al. (2005) noted that medical professionals receive "inadequate training in this important area, and their performance on this task remains less than ideal" (p. 652).

Autopsies and other confirmatory analyses can provide clarity about the cause of death where this is uncertain. While generally under-studied for people with intellectual disability, Kiani et al. (2014) suggest that autopsies were ordered less frequently for people with intellectual disability than those without intellectual disability for deaths from probable sudden unexpected death in epilepsy.

4 | DISCUSSION

A significant impact of inaccuracies and inconsistencies in MCCD for people with intellectual disability is the ability of researchers to accurately report the cause of death of people with intellectual disability. Tyrer and McGrother (2009) caution that "identifying people with ID from death certificates alone may not be possible" (p. 899), and Hosking et al. (2016) report that data solely taken

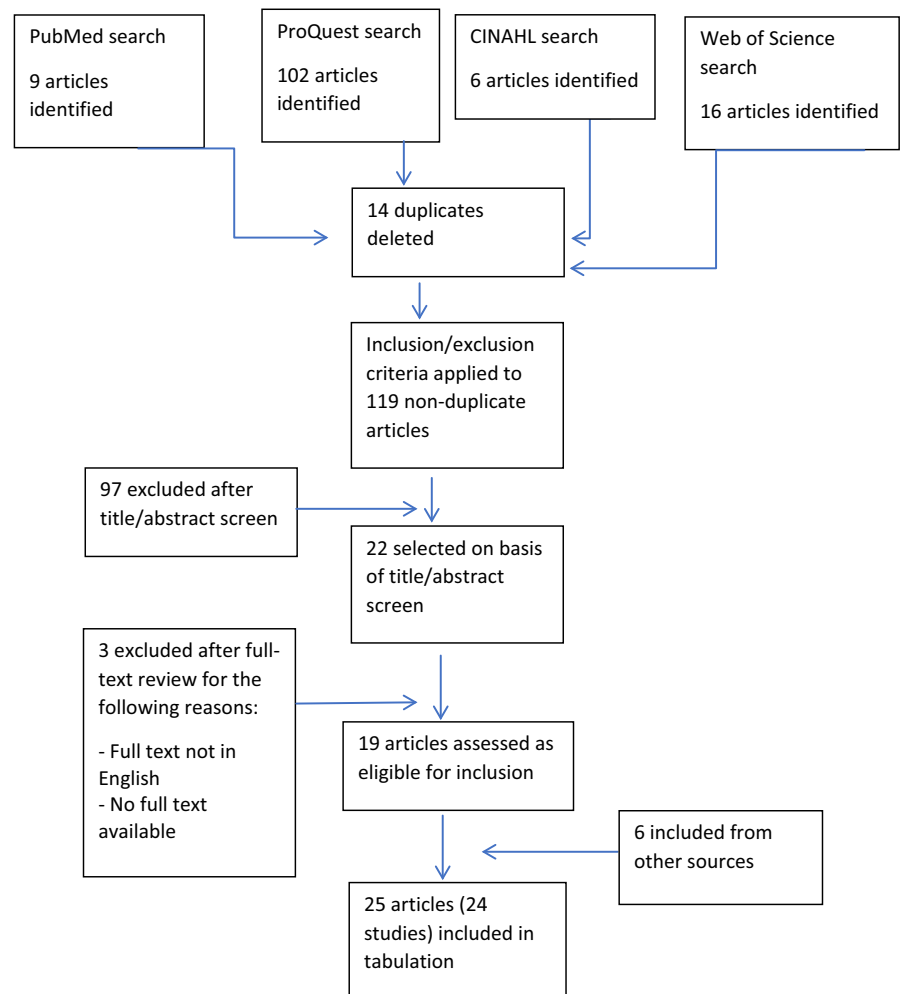


FIGURE 1 Results of search

from MCCD are “inadequate for understanding the mortality experience” of people with intellectual disability (p. 1483). Research is therefore complicated by the need to take account of the reliability of MCCD, with researchers having to link with other, often difficult to access data sets or to adapt their research methods. Trollor et al. (2017), for example, modified their data to create an “intellectual disability revised” group (p. 3), changing the underlying cause of death if it was a code for the aetiology of the person’s intellectual disability. Such revisions to research methods can increase the time and complexity of mortality studies, but more importantly can reduce the comparability of the findings in relation to other studies or population groups. This can have a potentially detrimental impact on health policy, which in the absence of easily available data about people with intellectual disability in national vital statistics relies upon primary research for information on mortality and morbidity trends, disease trajectories and the impact of intervention programmes (Landes & Peek, 2013).

A common theme across the reviewed literature is a focus on mortality of people with moderate, severe or profound intellectual disability. This could lead to a misunderstanding of the results of mortality studies; for example, it is less likely that people with mild intellectual disability would have intellectual disability mentioned on the MCCD. Although this was not specifically mentioned in the literature that was reviewed, a potentially confounding factor is the marking on MCCD of easily recognizable syndromes. The presence of Down’s syndrome, for example, is easily recognized and so may be more likely to be included on a MCCD; conversely, a person with subtle features of Foetal Alcohol Spectrum Disorder, or a non-syndromal cause of intellectual disability, may be less likely to have this recorded on the MCCD. Related to this is the erroneous inclusion of intellectual disability itself as a cause of death, which is not just a recent change in professional guidance; Baird and Sadovnick noted in 1988 “It is hard to envision how mental retardation in itself could cause death” (p. 243).

The literature reviewed includes an international spectrum of mortality studies, but the studies are largely confined to Western countries. While there are apparent variances in coding conventions between different countries, all follow WHO guidelines and the structure of ICD coding. This review aimed to draw overarching themes in relation to MCCD for people with intellectual disability, rather than the specific differences between different countries in relation to coding causes of death of people with intellectual disability, but this could be a productive area of future research.

5 | CONCLUSION

The prevalence of concerns about the accuracy and reliability of MCCD for people with intellectual disability raises questions about the validity of mortality data based on MCCD. One solution may be the provision of better informed guidance and training for those completing MCCD for people with intellectual disability (Hosking et al., 2016). A key focus of guidance could be ensuring

that intellectual disability or associated conditions are included on the MCCD not in Part 1 as part of the sequence of the death, but in Part 2 as a significant condition. Tyrer and McGrother (2009) also support the proposal by Hollins et al. (1998) that what is needed is a “place on the certificate to write down other conditions present at death, which are not related to the death” (p. 151). This would be advantageous in enabling the MCCD to be a useful tool for measuring mortality rates of people with intellectual disability without distorting the data and would support uniformity between countries. The organization to take a leading role in standardizing the guidance is the WHO. With the forthcoming ICD-11 revision and associated guidance, now is a critical period and a window for the WHO to act upon the findings of this and other studies.

CONFLICT OF INTEREST

None.

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REFERENCES

- Antini, C., Rajs, D., Munoz-Quezada, M. T., Mondaca, B. A., & Heiss, G. (2015). Reliability of cause of death coding: An international comparison. *Cadernos de Saude Publica*, 31(7), 1473–1482. <https://doi.org/10.1590/0102-311x00099814>
- Arvio, M., Salokivi, T., Tiitinen, A., & Haataja, L. (2016). Mortality in individuals with intellectual disabilities in Finland. *Brain and Behavior*, 6(2), e00431. <https://doi.org/10.1002/brb3.431>
- Baird, P. S., & Sadovnick, A. D. (1988). Causes of death to age 30 in Down syndrome. *American Journal of Human Genetics*, 43, 239–248.
- Bell, G. S., Gaitatzis, A., Johnson, A. L., & Sander, J. W. (2004). Predictive value of death certification in the case ascertainment of epilepsy. *Journal of Neurology, Neurosurgery & Psychiatry*, 75(12), 1756. <https://doi.org/10.1136/jnnp.2003.029918>
- Bugeja, L., Clapperton, A. J., Killian, J. J., Stephan, K. L., & Ozanne-Smith, J. (2010). Reliability of ICD-10 external cause of death codes in the National Coroners Information System. *Health Information Management Journal*, 39(3), 16–26. <https://doi.org/10.1177/183335831003900303>
- Cheng, T. J., Lu, T. H., & Kawachi, I. (2012). State differences in the reporting of diabetes-related incorrect cause-of-death causal sequences on death certificates. *Diabetes Care*, 35(7), 1572–1574. <https://doi.org/10.2337/dc11-2156>
- Dieckmann, F., Giovis, C., & Offergeld, J. (2015). The life expectancy of people with intellectual disabilities in Germany. *Journal of Applied Research in Intellectual Disabilities*, 28(5), 373–382. <https://doi.org/10.1111/jar.12193>
- Disability Rights Commission (2006). *Equal treatment: Closing the gap*. London: Disability Rights Commission.
- Florio, T., & Trollor, J. (2015). Mortality among a cohort of persons with an intellectual disability in New South Wales, Australia. *Journal of Applied Research in Intellectual Disabilities*, 28(5), 383–393. <https://doi.org/10.1111/jar.12190>
- Glover, G., & Ayub, M. (2010). How people with learning disabilities die. *Learning Disabilities Observatory*, 1–55. Retrieved from <https://>

- www.improvinghealthandlives.org.uk/uploads/doc/vid_9033_IHAL2010-06%20Mortality.pdf
- Goldman, E. S., Urbano, C. R., & Hodapp, M. R. (2011). Determining the amount, timing and causes of mortality among infants with Down syndrome. *Journal of Intellectual Disability Research*, 55(1), 85–94. <https://doi.org/10.1111/j.1365-2788.2010.01349.x>
- Haghighi, M. H., Dehghani, M., Teshizi, S. H., & Mahmoodi, H. (2014). Impact of documentation errors on accuracy of cause of death coding in an educational hospital in Southern Iran. *Health Information Management Journal*, 43(2), 35–42.
- Harriss, L. R., Ajani, A. E., Hunt, D., Shaw, J., Chambers, B., Dewey, H., ... Tonkin, A. (2011). Accuracy of national mortality codes in identifying adjudicated cardiovascular deaths. *Australian and New Zealand Journal of Public Health*, 35(5), 466–476. <https://doi.org/10.1111/j.1753-6405.2011.00739.x>
- Heslop, P., Blair, P. S., Fleming, P., Hoghton, M., Marriott, A., & Russ, L. (2014). The confidential inquiry into premature deaths of people with intellectual disabilities in the UK: A population-based study. *Lancet*, 383(9920), 889–895. [https://doi.org/10.1016/s0140-6736\(13\)62026-7](https://doi.org/10.1016/s0140-6736(13)62026-7)
- Heslop, P., & Glover, G. (2015). Mortality of people with intellectual disabilities in England: A comparison of data from existing sources. *Journal of Applied Research in Intellectual Disabilities*, 28(5), 414–422. <https://doi.org/10.1111/jar.12192>
- Heslop, P., Lauer, E., & Hoghton, M. (2015). Mortality in people with intellectual disabilities. *Journal of Applied Research in Intellectual Disabilities*, 28, 367–372. <https://doi.org/10.1111/jar.12196>
- Hogg, J., Juhlberg, K., & Lambe, L. (2007). Policy, service pathways and mortality: A 10-year longitudinal study of people with profound intellectual and multiple disabilities. *Journal of Intellectual Disability Research*, 51(5), 366–376. <https://doi.org/10.1111/j.1365-2788.2006.00884.x>
- Hollins, S., Attard, M. T., von Fraunhofer, N., McGuigan, S., & Sedgwick, P. (1998). Mortality in people with learning disability: Risks, causes, and death certification findings in London. *Developmental Medicine and Child Neurology*, 40(1), 50–56.
- Hosking, F. J., Carey, I. M., Shah, S. M., Harris, T., DeWilde, S., Beighton, C., & Cook, D. G. (2016). Mortality among adults with intellectual disability in England: Comparisons with the general population. *American Journal of Public Health*, 106(8), 1483–1490. <https://doi.org/10.2105/AJPH.2016.303240>
- Jha, P. (2012). Counting the dead is one of the world's best investments to reduce premature mortality. *Hypothesis*, 10, e3. Retrieved from <http://www.hypothesisjournal.com/?p=1301>
- Jopp, D. A., & Keys, C. B. (2001). Diagnostic overshadowing reviewed and reconsidered. *American Journal of Mental Retardation*, 106(5), 416–433. [https://doi.org/10.1352/0895-8017\(2001\)106<0416:dorar>2.0.co;2](https://doi.org/10.1352/0895-8017(2001)106<0416:dorar>2.0.co;2)
- Katsakiori, P. F., Panagiotopoulou, E. C., Sakellaropoulos, G. C., Papazafiropoulou, A., & Kardara, M. (2007). Errors in death certificates in a rural area of Greece. *Rural and Remote Health*, 7(4), 822.
- Kiani, R., Tyrer, F., Jesu, A., Bhaumik, S., Gangavati, S., Walker, G., ... Barrett, M. (2014). Mortality from sudden unexpected death in epilepsy (SUDEP) in a cohort of adults with intellectual disability. *Journal of Intellectual Disability Research*, 58(6), 508–520. <https://doi.org/10.1111/jir.12047>
- Kuijpers, C. C. H. J., Fronczek, J., van de Goot, F. R. W., Niessen, H. W. M., van Diest, P. J., & Jiwa, M. (2014). The value of autopsies in the era of high-tech medicine: Discrepant findings persist. *Journal of Clinical Pathology*, 67, 512–519. <https://doi.org/10.1136/jclinpath-2013-202122>
- Lakhan, R., & Kishore, T. M. (2016). Mortality in people with Intellectual Disability in India: Correlates of age and settings. *Life Span and Disability*, 19(1), 45–56.
- Landes, S. D., & Peek, C. W. (2013). Death by mental retardation? The influence of ambiguity on death certificate coding error for adults with intellectual disability. *Journal of Intellectual Disability Research*, 57(12), 1183–1190. <https://doi.org/10.1111/j.1365-2788.2012.01614.x>
- Lauer, E., & McCallion, P. (2015). Mortality of people with intellectual and developmental disabilities from select US state disability service systems and medical claims data. *Journal of Applied Research in Intellectual Disabilities*, 28(5), 394–405. <https://doi.org/10.1111/jar.12191>
- Mahdavi, A., Sedghi, S., Sadoghi, F., & Fard Azar, F. E. (2015). Assessing the awareness of agents involved in issuance of death certificates about death registration rules in Iran. *Global Journal of Health Science*, 7(5), 371–379. <https://doi.org/10.5539/gjhs.v7n5p371>
- McCarron, M., Carroll, R., Kelly, C., & McCallion, P. (2015). Mortality rates in the general Irish population compared to those with an intellectual disability from 2003 to 2012. *Journal of Applied Research in Intellectual Disabilities*, 28(5), 406–413. <https://doi.org/10.1111/jar.12194>
- McCarthy, J., & O'Hara, J. (2011). Ill-health and intellectual disabilities. *Current Opinion in Psychiatry*, 24(5), 382–386. <https://doi.org/10.1097/YCO.0b013e3283476b21>
- McKenzie, K., Chen, L., & Walker, S. M. (2009). Correlates of undefined cause of injury coded mortality data in Australia. *Health Information Management Journal*, 38(1), 8–14. <https://doi.org/10.1177/183335830903800102>
- Ministry of Justice (2016). *Coroners Statistics Annual 2016 England and Wales*. Retrieved from: https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/613556/coroners-statistics-2016.pdf
- Miodrag, N., Silverberg, S. E., Urbano, R. C., & Hodapp, R. M. (2013). Deaths among children, adolescents, and young adults with down syndrome. *Journal of Applied Research in Intellectual Disabilities*, 26, 207–214. <https://doi.org/10.1111/jar.12023>
- NCEPOD (National Confidential Enquiry into Patient Outcome and Death) (2006). *The Coroner's autopsy: Do we deserve better?* London, UK: NCEPOD. Retrieved from: <http://www.ncepod.org.uk/2006Report/Downloads/Coronial%20Autopsy%20Report%202006.pdf>
- Ouellette-Kuntz, H. (2005). Understanding health disparities and inequities faced by individuals with intellectual disabilities. *Journal of Applied Research in Intellectual Disabilities*, 18(2), 113–121. <https://doi.org/10.1111/j.1468-3148.2005.00240.x>
- Ouellette-Kuntz, H., Shooshtari, S., Balogh, R., & Martens, P. (2015). Understanding information about mortality among people with intellectual and developmental disabilities in Canada. *Journal of Applied Research in Intellectual Disabilities*, 28(5), 423–435. <https://doi.org/10.1111/jar.12195>
- Rahu, K., Palo, E., & Rahu, M. (2011). Diminishing trend in alcohol poisoning mortality in Estonia: Reality or coding peculiarity? *Alcohol and Alcoholism*, 46(4), 485–489. <https://doi.org/10.1093/alcalc/agr046>
- Raitasuo, J., Molsa, P., Raitasuo, S., & Mattila, K. (1997). Deaths among the intellectually disabled: A retrospective study. *Journal of Applied Research in Intellectual Disabilities*, 10(4), 280–288. <https://doi.org/10.1111/j.1468-3148.1997.tb00023.x>
- Rampatige, R., Gamage, S., Peiris, S., & Lopez, A. D. (2013). Assessing the reliability of causes of death reported by the Vital Registration System in Sri Lanka: Medical records review in Colombo. *Health Information Management Journal*, 42(3), 20–28. <https://doi.org/10.12826/18333575.2013.0009.Rampatige>
- Reiss, S., Levitan, G. W., & Szyszko, J. (1982). Emotional disturbance and mental retardation: Diagnostic overshadowing. *American Journal of Mental Deficiency*, 86(6), 567–574.
- Shojania, K. G., & Burton, E. C. (2008). The vanishing nonforensic autopsy. *The New England Journal of Medicine*, 358(9), 873–875. <https://doi.org/10.1056/NEJMp0707996>
- Swain, G. R., Ward, G. K., & Hartlaub, P. P. (2005). Death certificates: Let's get it right. *American Family Physician*, 71(4), 652–656.
- Trollor, J., Srasuekul, P., Xu, H., & Howlett, S. (2017). Cause of death and potentially avoidable deaths in Australian adults with intellectual

- disability using retrospective linked data. *British Medical Journal Open*, 7(2).
- Tyrer, F., & McGrother, C. (2009). Cause-specific mortality and death certificate reporting in adults with moderate to profound intellectual disability. *Journal of Intellectual Disability Research*, 53(11), 898–904. <https://doi.org/10.1111/j.1365-2788.2009.01201.x>
- Tyrer, F., Smith, K. L., & McGrother, W. C. (2007). Mortality in adults with moderate to profound intellectual disability: A population-based study. *Journal of Intellectual Disability Research*, 51(7), 520–527. <https://doi.org/10.1111/j.1365-2788.2006.00918.x>
- World Health Organisation (2010). *International Statistical Classification of Diseases and Related Health Problems 10th Revision Volume 2 Instruction manual 2010 Edition*. Retrieved from http://www.who.int/classifications/icd/ICD10Volume2_en_2010.pdf
- World Health Organisation (2014). *Civil registration: why counting births and deaths is important*. Retrieved from <http://www.who.int/mediacentre/factsheets/fs324/en/>
- Yang, Q. H., Rasmussen, S. A., & Friedman, J. M. (2002). Mortality associated with Down's syndrome in the USA from 1983 to 1997: A population-based study. *Lancet*, 359(9311), 1019–1025. [https://doi.org/10.1016/s0140-6736\(02\)08092-3](https://doi.org/10.1016/s0140-6736(02)08092-3)
- Ylijoki-Sorensen, S., Sajantila, A., Lalu, K., Boggild, H., Boldsen, J. L., & Boel, L. W. (2014). Coding ill-defined and unknown cause of death is 13 times more frequent in Denmark than in Finland. *Forensic Science International*, 244, 289–294. <https://doi.org/10.1016/j.forsciint.2014.09.016>

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